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Case report

Evolution of 'Dementia Infantilis to Childhood Disintegrative Disorder, or Autism Spectrum Disorder': A case report

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Abstract

Childhood disintegrative disorder (CDD), rare clinical syndrome characterized by at least two years of normal development, followed by distinct regression of developmental and behavioral functioning before ten years of age. CDD is a condition that no longer officially exists, been eliminated and absorbed into the larger category of autism spectrum disorder with the release of the 5th edition of the Diagnostic and Statistical Manual of Mental Disorders (DSM-5). Here we report the case of 12 years old male who presented normal development till 4 years of age followed by gradual deterioration in his communication, motor and social skills with behavioral disturbances. The aim of our case report is to increase awareness of this rare illness, due to diagnostic dilemma, especially when no identifiable organic cause is elicited so early diagnosis and prompt treatment required.

Key words: Autism spectrum disorder, Heller's syndrome, dementia infantilis, pervasive developmental disorder.

Introduction

Childhood disintegrative disorder (CDD) is characterized by an apparently normal development for at least the first two to three years after birth, as manifested by the presence of age-appropriate verbal and nonverbal communication, social relationships, play, and adaptive behavior. Then patient usually presented with severe reversal in language, social function and motor and cognitive skills [1, 2]. It is associated with behavioral, affective symptoms such as fear, over activity and rarely hallucinations. The prevalence of CDD is 1 in 100,000 children and it is common in boys than girls, with a ratio 8:1 respectively. CDD is also sometimes associated with seizures.

CDD has been recognized for century ago in 1908 by an Austrian educator, Theodore Heller so it is also called Heller's syndrome. It is complex disorder over the years with various names as 'progressive disintegrative psychosis', 'disintegrative psychosis', 'pervasive disintegrative disorder' [3], as 'other childhood disintegrative disorder' (other CDD) in International Classification of Diseases-10 criteria (ICD 10) [4]. CDD, Autistic disorder, Rett's syndrome as well as Asperger's syndrome are under category of 'Autistic spectrum disorder (ASD)' under DSM-5 [5]. In 2013, however, the release of the DSM-5 made CDD's official recognition in the psychiatric field short-lived. ASD replaces the group of disorders called pervasive developmental disorders including autistic disorder, childhood disintegrative disorder, Asperger's disorder, and pervasive developmental disorder not otherwise specified (PDD-NOS).

Heller called it as 'dementia infantilis', as it similar to senile dementia due to developmental regression [6]. It is also called 'disintegrative psychosis of childhood' and described as normal or near normal development for first few years of life, onset started after three and a half years, and severe disintegration such as a deterioration of emotions, behavior, and relationships and often a

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loss of speech and other functions [7]. It is also named as 'progressive disintegrative psychosis of childhood' to differentiate from psychiatric illnesses [8].

There is no specific pharmacological treatment in ASD, various psychotropic like risperidone olanzapine and clozapine have shown effectiveness [9,10] and trial of benzodiazepine and stimulant are helpful to control aggression. Psychosocial treatment is also beneficial such as behavior modification, psycho-education to child and parents, speech and language therapy, social skill development and support group. Behavioral therapy should be focused on (a) development of a regular routine, (b) structured class room training, aiming at learning new material and maintenance of acquired learning, (c) positive reinforcements to teach self-care skills, (d) speech therapy and sign language teaching and (e) behavioral techniques to encourage interpersonal interactions. Parental counseling and supportive psychotherapy are helpful in reducing the parental anxiety and guilt, and ensuring their active involvement in therapy. Here we present the case of a boy who presented normal development till 4 years of age followed by gradual deterioration in his communication, motor and social skills with significant behavioral disturbances.

Case history

A 12-year-old boy brought to our emergency department by his parents with complaints of irritable behavior, not able to speak, decreased interaction with family members and siblings. He was making problems in routine daily activities since age of eight years. He was born with full term cesarean section delivery due to prolonged labor in parents of consanguineous marriage. His further history revealed that birth milestone achieved normal and patient was absolutely normal till age of four years. There were no significant antenatal, perinatal or post- natal complications and he was fully immunized. Patient had normal developmental milestone and acquired age appropriate social and communication skills until the age of four years. At the age of four when

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his family visited to a farm house, then during playing he went beside desert and missed his way to come back home. Later, after searching he was found crying in dirty clothes. Initially parents noticed that at times he would appear very fearful, and on enquiry would not be able to give any reason for it. He gradually stopped performing tasks. His parents realized that the patient was not interested with his siblings, stopped going for playing. He responded to questions occasionally with nod, and his speech deteriorated with monosyllabic and unintelligible. He was also noticed that auto-regression in the form of biting himself or banging his head against the wall showing his annoyance. He also observed physically aggressive towards other people; throwing household things, also noticed he would remain occupied in his own world doing stereotypic behavior as grinding teeth and winging his fingers periodically. There was also marked deterioration in his motor skills such as he was unable to grasp things.

Patient was taken to private hospital during the onset of illness. He was investigated thoroughly with computed tomography scan, electroencephalogram but all reports were unremarkable and took some medications, but detail were not available. From that time no improvement was perceived and he was referred to our center for second opinion.

On mental state examination his eye to eye contact was poor and he would do aimless activities without showing any interest in his surroundings. He was hardly speaking with monosyllables, restless and exhibiting stereotypic wringing of fingers.

On examination there was no any evidence of systemic or neurological abnormalities. His vitals were within normal range. All routine investigations like complete blood count, metabolic work up includes blood sugar, albumin, liver function test, fasting blood test for triglyceride and cholesterol and vitamin B12 level and kidney, thyroid function test was within normal limit.

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Electroencephalogram and computed tomography scan were unremarkable. There is no family history of psychiatric illness or any history of mental retardation.

Patient was started on tablet risperidone 1mg twice per day and gradually increased up to 3 mg per day, and over the next three weeks from which he showed minimal improvement. Patient has been referred to a higher center for rehabilitation program includes speech, language therapy and educational support. It helps the patient lead a more functional and productive life and improve his cognitive abilities.

Patient has been given psycho-education and supportive psychotherapy to relieve care givers burden. Patient was following up every month after three months trivial improvement was found. On subsequent follow up after six months his social interaction improved and he started play with other children at home and also shown some improvement in communication.

Discussion

In this case, normal developmental milestones till the age of four years, then gradual loss of communication skills, cognitive and social skills, marked regression, loss of ability to relate to people and absence of any other neurological deficit were the points in favor of diagnosis of Autism Spectrum Disorder.

There was no history of any delusions, hallucinations, and disorganized speech, so it seems less chance of childhood schizophrenia. CDD, a part of a larger category called ASD, is a neuropsychiatric neurodevelopmental disorder. Unlike autism, children with CDD show severe regression after several years of normal development and more dramatic loss of skills than a child with autism does.

Pharmacotherapy with atypical antipsychotics has shown varying degrees of effectiveness in controlling behavioral problems in PDD patient; therefore, they are used in patients with ASD as

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well. Benzodiazepine is helpful to decrease aggression as well improvement of auto-regression. Supportive psychotherapy and psycho-education for families play important role to reduce their isolation and frustration. A comprehensive rehabilitation program includes speech therapy, language therapy and occupational therapy helpful for more functional and productive life. Rehabilitation plan targets child's communication skill with speech and language therapy, social interaction skill with counseling and simulated scenarios and motor function skill with occupational and physical therapy.

The patient was diagnosed as Autism Spectrum Disorder as per DSM-5 diagnostic criteria [4] and Other Childhood Disintegrative Disorder as per ICD-10 diagnostic criteria, the new ASD diagnostic criteria describe two primary sets of symptoms: 1) social communication and social interaction deficit, and 2) restrictive and repetitive behavior patterns. Each of these symptom sets (social and behavioral) will include three severity levels to identify the supportive services that are required. These three severity levels are: 1) requiring support, 2) requiring substantial support, and 3) requiring very substantial support. The DSM-5 describes each of these severity levels, with examples, to aid clinicians in making these determinations.

In DSM-5 it has been unclear whether individual with CDD have a late-onset, regressive form of autism, or whether this category captures a truly distinct condition that affects certain children. In ICD-10 they found that the children with CDD had accumulated more skills than the children with autism prior to regression and had worse speech and intellectual disabilities, so they concluded that CDD merits a separate diagnosis from the late-onset autism. That's why controversy in the autism field about whether CDD really a distinct entity, worthy of its own diagnostic category and need a high degree of research scrutiny.

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To conclude, evolution (changes in different names) comes over the years with various names of CDD. Actually, a primary reason for this change was that research demonstrated wide variability among clinicians in applying the diagnostic criteria for this group of disorders. At present according to DSM-5 criteria it's called Autism Spectrum Disorder and ICD-10 called other childhood disintegrative disorder.

Developmental milestone history and screening is important for early detection and awareness of the disorder. In this type of illness children always need support with the activities of daily living and long-term care facility. However, early detection of illness and early comprehensive intervention can help improvement and alleviate deterioration for children even though there is poor prognosis of this illness. More research and pharmacological trials are needed to improve this illness in coming years.

Conflict of interest: None declared

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